

PEDIATRIC CARDIOLOGY

Recognition of Abnormal Connections of Coronary Arteries With the Use of Doppler Color Flow Mapping

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Anomalous connection of a coronary artery to a ventricle or pulmonary artery causes shunting of blood from the coronary circuit and may produce myocardial ischemia. Such a coronary anomaly may occur in isolation or with other defects. Doppler color flow mapping and two-dimensional echocardiography were used to diagnose anomalous coronary connections in 13 patients, 1 day to 7 years of age, over a 1 year period. The diagnoses were anomalous origin of the left coronary artery from the pulmonary trunk in five patients, a coronary artery to left ventricle fistula or coronary artery to pulmonary artery fistula in four patients with other complex defects, right ventricular sinusoids in

two patients with pulmonary atresia and intact ventricular septum and an isolated coronary artery fistula in two patients.

In all cases, the abnormal coronary connection was recognized on the basis of an abnormal, continuous or to and fro flow pattern in the fistula and its connections as demonstrated by scanning in multiple views with Doppler color flow mapping. The low spatial resolution of Doppler color flow mapping limits the anatomic detail available; nonetheless, it is a significant advance in the noninvasive diagnosis of abnormal coronary connections.

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Anomalous connection of a coronary artery to a heart chamber or a pulmonary artery causes shunting of blood from the coronary circuit. This occurrence may result in myocardial ischemia or infarction and a left to right shunt. Such an anomalous coronary connection may occur in isolation or in association with other heart defects. For example, anomalous origin of the left coronary artery from the pulmonary trunk is usually an isolated lesion and generally results in ischemic, dilated cardiomyopathy with or without infarction (1-3). Detection of anomalous left coronary artery by two-dimensional echocardiography and pulsed Doppler cardiography is possible but difficult and unreliable (4-6).

Coronary artery fistulas may occur in isolation or along with other defects such as ventricular outlet atresia (7-10) or stenosis. Many case reports (11-18) document the ability of two-dimensional echocardiography, with or without contrast injection, and pulsed Doppler echocardiography to detect coronary artery fistulas. More recently, Doppler color flow mapping was used to diagnose such a fistula (19). In nearly

all reported cases, however, the fistula was suspected on clinical grounds and was an isolated lesion. Coronary artery fistulas associated with complex congenital heart defects rarely produce findings that arouse clinical suspicion and therefore may escape detection by any modality.

We have found Doppler color flow mapping to be extremely valuable in detecting abnormal coronary artery connections, especially in association with other complex heart defects. We describe 13 patients in whom an abnormal coronary artery connection was detected by Doppler color flow mapping.

Methods

Selection of patients. The records of the noninvasive laboratory of the Department of Cardiology in this institution were searched for the calendar year 1987 for any patient diagnosed with an abnormal coronary connection, including anomalous origin of the left coronary artery from the pulmonary trunk, a coronary artery fistula or a coronary sinusoid in pulmonary atresia with intact ventricular septum. Similarly, the records of the catheterization laboratory of the Department of Cardiology and the surgical reports of the Division of Cardiovascular Surgery were searched for any patient with an abnormal coronary connection detected by angiography or at surgery during the same period. The

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echocardiogram, echocardiographic report and clinical records were reviewed for all patients identified.

Two-dimensional echocardiography and Doppler color flow mapping. The procedures were performed with the use of a Hewlett-Packard 77020 cardiac imager with Doppler ultrasound. A 5.0 or 3.5 MHz transducer was used for each examination. Patients were examined supine or in a left or right decubitus position in subxiphoid, apical, parasternal and suprasternal notch views. In addition to imaging, Doppler color flow mapping was performed while the heart was scanned in all views. The low velocity setting (lowering the cutoff value for the high pass filter) was used frequently to enhance the amplitude of the signal from the coronary arteries. Sedation with chloral hydrate was used when necessary in infants and young children. All examinations were recorded on 1/2 in. (1.3 cm) video cassette tape and were reviewed before final interpretation in real time, slow motion and stop-frame modes. The examiner was aware of the findings on physical examination, electrocardiogram, chest X-ray film and any previous echocardiogram or angiogram.

Results

Study patients. During 1987 18 patients, aged 1 day to 7.5 years, were diagnosed as having an abnormal coronary connection by angiography or surgical observation, or both. The abnormal connection was detected in 13 patients by echocardiography and Doppler color flow mapping. In the remaining five patients with pulmonary atresia and intact ventricular septum, right ventricular sinusoids were not detected by echocardiography. However, Doppler color flow mapping was not performed in three of these five patients and was not used to scan the right ventricle in the other two. All abnormal coronary artery connections diagnosed by echocardiography and Doppler color flow mapping were subsequently confirmed at angiography or surgery, or both.

Anomalous origin of the left coronary artery from the pulmonary trunk. This anomaly was present in five patients, aged 2 months to 7 years. In three of them, the diagnosis was made prospectively by two-dimensional echocardiography and Doppler color flow mapping (Fig. 1a and b). The fourth infant had undergone cardiac catheterization at another institution before referral to this hospital. However, the aortic root angiogram failed to demonstrate the anomalous origin of the left coronary artery. The diagnosis was made by echocardiography and Doppler color flow mapping, which showed that the left coronary artery arose from the pulmonary trunk distally, near the origin of the right pulmonary artery. The fifth patient, a 7 year old girl, was initially evaluated at this hospital in 1980 because of dilated cardiomyopathy. At that time, an echocardiogram (without Doppler ultrasound) and a left ventricular angiogram were interpreted as showing normal origin of the left coronary artery.

During a recent echocardiogram for follow-up of mitral regurgitation and evaluation of left ventricular function, both coronary arteries were noted to be dilated and the coronary artery anatomy was again investigated. Doppler color flow mapping was instrumental in documenting the pulmonary origin of the left coronary artery by demonstrating the flow jet from the coronary ostium into the pulmonary trunk. The diagnosis was confirmed in all five patients at surgery.

The exact site of connection of the left coronary artery with the pulmonary trunk was correctly determined in four of the five cases by echocardiography and Doppler color flow mapping. In the fifth case, the lateral part of an intramural segment of the left coronary artery was interpreted as the site of connection with the pulmonary trunk because of spillover of the color signal into the pulmonary trunk. Review of the examination after surgical delineation of the anatomy showed the intramural segment and the real orifice medial and near the pulmonary valve.

Coronary fistula in association with a complex congenital heart defect. This anomaly was diagnosed prospectively by echocardiography and Doppler color flow mapping in four infants on the first day of life. One infant with tetralogy of Fallot had a fistula between the left coronary artery and the mid right pulmonary artery. The right pulmonary artery was in continuity with the main pulmonary artery and right ventricular outflow tract. The left pulmonary artery was isolated, receiving blood only from the ductus arteriosus. The findings that led to the diagnosis were mild dilation of the left coronary artery and continuous flow in the distal right pulmonary artery but phasic flow from the right ventricular outflow tract in the proximal right pulmonary artery. Clockwise rotation of the transducer from the parasternal short-axis view displayed the coronary artery and its connection with the right pulmonary artery.

Another infant with tetralogy of Fallot, pulmonary atresia and discontinuous branch pulmonary arteries had a large fistula between the proximal right coronary artery and the proximal right pulmonary artery. Doppler color flow mapping in a parasternal short-axis view demonstrated continuous flow from the right coronary artery through the fistula and into the pulmonary artery (Fig. 1c and d). The coronary artery to pulmonary artery fistula was not demonstrated at the initial catheterization when injections were made into the left ventricle, ascending aorta and descending aorta. A selective right coronary artery injection at a subsequent catheterization showed the fistula.

The other two infants had severe left ventricular hypoplasia with atresia of the left ventricular outflow tract, an intact ventricular septum and a patent mitral valve. In one case the diagnosis was d-transposition of the great arteries with valvular pulmonary atresia. A fistula between the hypoplastic left ventricle and the left anterior descending coronary artery through a septal perforating branch was detected by

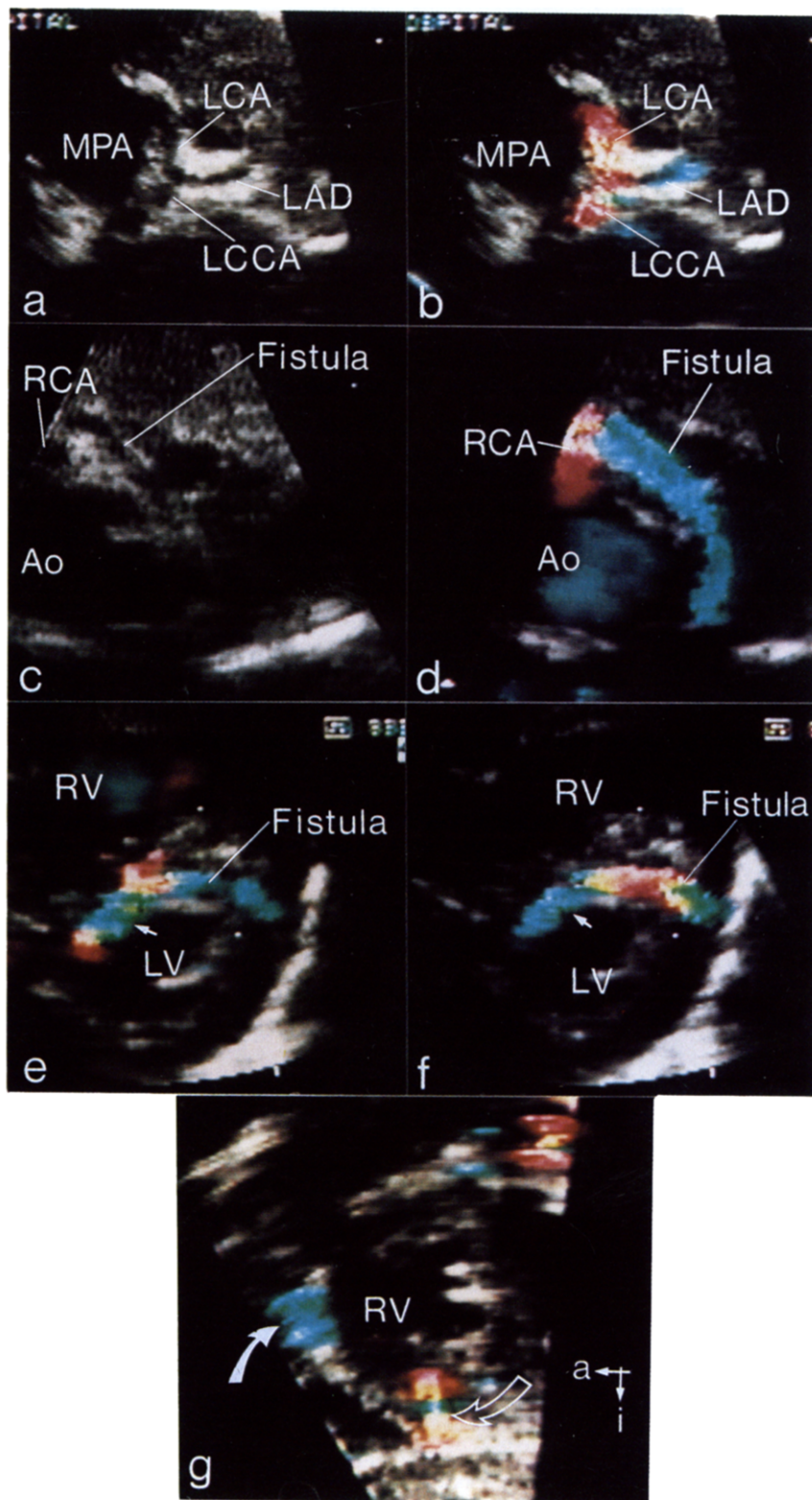


Figure 1. Doppler color flow mapping in four cases of abnormal coronary artery connection. **a**, Parasternal short-axis view in a patient with anomalous left coronary artery (LCA). The left coronary artery with branches can be seen connecting with the lateral aspect of the main pulmonary artery (MPA). **b**, In the same patient, a similar view on Doppler color flow mapping shows the flow stream from the dilated left coronary artery (LCA) into the main pulmonary artery (MPA). Flow can be seen in the branch coronary arteries as well (left circumflex artery [LCCA] and left anterior descending [LAD] artery). **c**, A fistula from the right coronary artery (RCA) to the right pulmonary artery in a patient with tetralogy of Fallot, pulmonary atresia and discontinuous branch pulmonary arteries. The fistula, passing to the left of the aortic root (AO), is difficult to see by imaging alone. **d**, In the same patient, the fistula is readily apparent by Doppler color flow mapping. Flow in the right coronary artery (RCA) (red) is toward the transducer, whereas flow in the fistula (blue) is away from the transducer. **e**, Doppler color flow mapping in the parasternal short-axis view demonstrating a fistula between the hypoplastic left ventricle (LV) and the left coronary artery system. In systole, flow is from the left ventricle to the coronary artery. Flow is toward the transducer in the rightward end of the vessel and away in the leftward end. The blue color surrounded by red in the rightward end is due to aliasing. The **arrow** marks the mouth of the fistula into the left ventricle. **f**, In the same patient in diastole flow reverses from the coronary artery to the left ventricle. Now flow is toward the transducer in the leftward end and away in the rightward end of the fistula. Now aliasing is noted at the leftward end of the vessel. RV = right ventricle. **g**, In this patient, Doppler color flow mapping in the subxiphoid short-axis view illustrates blood flow from the right ventricular (RV) cavity into a sinusoid on the diaphragmatic free wall of the right ventricle (**open arrow**). A red flow map indicates flow toward the transducer. Another part of the sinusoid is seen in the anterior free wall (**solid arrow**). The blue flow map indicates flow away from the transducer. This sinusoid, like most, is too tortuous to be displayed in a single imaging plane. a = anterior; i = inferior.

scanning with Doppler color flow mapping in parasternal and subxiphoid short-axis views (Fig. 1e and f). The ostium and proximal left coronary artery could not be imaged, suggesting stenosis or atresia. An aortic root injection failed to fill

the left coronary artery, supporting the echocardiographic impression. In the other patient the diagnoses included dextrocardia, double outlet right ventricle with l-ventricular loop (ventricular inversion) and pulmonary stenosis. The

fistula connecting the hypoplastic left ventricle with the posterior descending artery was detected by scanning with Doppler color flow mapping in apical four chamber and subxiphoid short-axis views. In the latter two cases, a strikingly abnormal flow pattern of nearly continuous to and fro flow was noted in the involved coronary artery. The coronary arteries were only mildly dilated.

Right ventricular sinusoids. These were detected in two patients, 1 day and 2 years old, respectively, with pulmonary atresia and intact ventricular septum. The sinusoids were diagnosed prospectively in the infant but had been demonstrated previously by angiography in the other child. In both cases, the abnormal coronary vessels were seen in the right ventricular septum or free wall by scanning with Doppler color flow mapping in subxiphoid, parasternal and apical views (Fig. 1g). However, details of coronary artery anatomy including coronary stenosis or atresia could not be demonstrated.

Seven other patients with pulmonary atresia and intact ventricular septum were examined during 1987. In two patients sinusoids were not detected by either Doppler color flow mapping or angiography. In the other five patients, one or more sinusoids were demonstrated by angiography but not by echocardiography. In three of these five patients, Doppler color flow mapping was not performed during the echocardiographic examination. In the other two patients, the Doppler color examination was limited to the mitral valve and pulmonary arteries without examination of the right ventricle.

Isolated coronary artery fistula. This defect was demonstrated by echocardiography and Doppler color flow mapping in two patients, aged 2 and 5 years, respectively. The fistula occurred between the left circumflex coronary artery and right atrium in one child and between the right coronary artery and right atrium in the other. The latter case had been diagnosed previously by angiography, whereas the former was diagnosed prospectively with echocardiography and Doppler color flow mapping. In both cases the fistula was best seen in a parasternal short-axis view. Doppler color flow mapping demonstrated a continuous flow pattern in the abnormal coronary artery and disturbed flow in the right atrium near the orifice of the fistula.

Discussion

Role of Doppler color mapping in diagnosis. Although uncommon, an abnormal connection of a coronary artery is a serious heart defect that often in itself produces symptoms or complicates management of other defects. These preliminary observations indicate that Doppler color flow mapping is a sensitive method for detecting such connections. However, it is possible that in some patients an abnormal coronary connection that was missed by echocardiography and Doppler color flow mapping went undetected by other

means as well. Most patients evaluated in the noninvasive laboratory do not undergo angiography or surgery, and these procedures also can fail to disclose an abnormal connection. Consequently, the actual sensitivity of the Doppler technique cannot be determined, but it appears to approach that of angiography. Of note, there were no false positive diagnoses.

In some cases, those with anomalous origin of the left coronary artery from the pulmonary trunk and isolated coronary artery fistula, the coronary anomaly was suspected clinically. Despite clinical suspicion, anomalous left coronary artery has proved to be extremely difficult to diagnose by echocardiography and pulsed Doppler ultrasound. In those cases where the left coronary artery has a nearly normal course, it often appears to connect with the left coronary sinus of the aorta due to false drop-out of the wall between the coronary artery and the aorta (4). Doppler color flow mapping demonstrates the abnormal flow from the coronary artery into the pulmonary artery (20) and, therefore, does not rely on imaging the thin wall between the coronary artery and the aorta for the diagnosis. Although the abnormal flow may be detected by pulsed Doppler ultrasound, it may also be missed despite an extensive search. Further, imaging the flow stream with Doppler color flow mapping more readily allows flow from a coronary artery to be distinguished from other unusual flow patterns due to swirling or reflection from the bifurcation.

In patients with associated defects the abnormal coronary connection was not suspected clinically. The clinical features of the associated defect usually obscured those that might be produced by the coronary abnormality. In our four cases of this type, the coronary arteries were only mildly dilated and the abnormal coronary connection might have been overlooked. Visualization of the abnormal flow pattern in the coronary artery or its connections by Doppler color flow mapping was the only clue to the diagnosis. It seems unlikely that the coronary fistula would have been detected in three of the patients without Doppler color flow mapping.

The clinical importance of these coronary anomalies was alluded to earlier. Timely diagnosis and surgical treatment of the anomalous left coronary artery usually results in virtually complete recovery of left ventricular function (3). If not interrupted, a fistula in association with another defect may be the route of continued shunting of blood from the coronary circulation even after repair of the associated defect. In cases with ventricular outlet obstruction the fistula is often a marker for a more generalized coronary arteriopathy.

Limitations of Doppler color flow mapping for defining coronary abnormalities. These limitations are related to the spatial resolution. The sample gates for Doppler color flow mapping are relatively large, resulting in poorer spatial resolution for flow mapping than for imaging. This result limits the anatomic detail that can be inferred from the flow map. For example, the number of orifices of the isolated

coronary fistula into the right atrium could not be determined. Also, the details of the right ventricular sinusoids, including the number of connections and which, if any, coronary artery was supplied by the sinusoid, could not be determined. Although the exact site of connection of the left coronary artery with the pulmonary trunk was determined in four of five patients, low spatial resolution of the Doppler color flow mapping led to an error in the fifth case as described.

The angle of interrogation seemed to have little effect on the amplitude of the flow map in the fistulous connection. However, flow in the more distal segments of the involved coronary artery was better imaged when the vessel was parallel to the Doppler beam.

Flow in curvilinear vessels may be toward the transducer at one end of the vessel and away at the other end (Fig. 1c and d). This occurrence results in a different color mapped at the two ends of the vessel. Because aliasing occurs at a low flow velocity with Doppler color flow mapping, color reversal is common in segments of vessels that are nearly parallel to the Doppler beam. This factor may be particularly confusing when trying to determine the direction of flow in a vessel. In such cases pulsed Doppler echocardiography may be useful.

Artifacts due to wall motion should not be confused with abnormal coronary flow because the latter tends to be nearly continuous and often disturbed, whereas the former is brief and intermittent and mapped as a pure color. Even lowering the cutoff value for the high pass filter does not produce artifacts that resemble abnormal coronary artery flow.

References

1. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk: its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. *Circulation* 1968;38:403-25.
2. Askenazi J, Nadas AS. Anomalous left coronary artery originating from the pulmonary artery: report on 15 cases. *Circulation* 1975;51:976-87.
3. Rein AJJT, Colan SD, Parness IA, Sanders SP. Regional and global left ventricular function in infants with anomalous origin of the left coronary artery from the pulmonary trunk: preoperative and postoperative assessment. *Circulation* 1987;75:115-23.
4. Robinson PJ, Sullivan ID, Kumpeng V, Anderson RH, Macartney FJ. Anomalous origin of the left coronary artery from the pulmonary trunk: potential for false negative diagnosis with cross sectional echocardiography. *Br Heart J* 1984;52:272-7.
5. Fisher EA, Sepehri B, Lendrum B, Luken J. Two-dimensional echocardiographic visualization of the left coronary artery in anomalous origin of the left coronary artery from the pulmonary artery. *Circulation* 1981;63:698-704.
6. Caldwell RL, Hurwitz RA, Girod DA, Weyman AE, Feigenbaum H. Two-dimensional echocardiographic differentiation of anomalous left coronary artery from congestive cardiomyopathy. *Am Heart J* 1983;106:710-6.
7. Sissman NJ, Abrams HL. Bidirectional shunting in a coronary artery-right ventricular fistula associated with pulmonary atresia and intact ventricular septum. *Circulation* 1965;32:582-8.
8. O'Connor WN, Cash JB, Cottrill CM, Johnson GL, Noonan JA. Ventriculocoronary connections in hypoplastic left hearts: an autopsy microscopic study. *Circulation* 1982;66:1078-86.
9. O'Connor WN, Cottrill CM, Johnson GL, Noonan JA, Todd EP. Pulmonary atresia with intact ventricular septum and ventriculocoronary communications: surgical significance. *Circulation* 1982;65:805-9.
10. Kasznica J, Ursell PC, Blanc WA, Gersony WM. Abnormalities of the coronary circulation in pulmonary atresia and intact ventricular septum. *Am Heart J* 1987;114:1415-20.
11. Reeder GS, Tajik AJ, Smith HC. Visualization of coronary artery fistula by two-dimensional echocardiography. *Mayo Clin Proc* 1980;55:185-9.
12. Kronzon I, Winer HE, Cohen M. Noninvasive diagnosis of left coronary arteriovenous fistula communicating with the right ventricle. *Am J Cardiol* 1982;49:1811-3.
13. Rodgers DM, Wolf NM, Barrett MJ, Zuckerman GL, Meister SG. Two-dimensional echocardiographic features of coronary arteriovenous fistula. *Am Heart J* 1982;104:872-4.
14. Satomi G, Endo M, Takao A, Nakamura K. A case of right coronary artery to left ventricle fistula: two-dimensional echocardiographic study. *Pediatr Cardiol* 1983;4:229-32.
15. Chen CC, Hwang B, Hsiung MC, et al. Recognition of coronary arterial fistula by Doppler 2-dimensional echocardiography. *Am J Cardiol* 1984;53:392-4.
16. Agatston AS, Chapman E, Hildner FJ, Samet P. Diagnosis of a right coronary artery-right atrial fistula using two-dimensional and Doppler echocardiography. *Am J Cardiol* 1984;54:238-9.
17. Miyatake K, Okamoto M, Kinoshita N, Fusejima K, Sakakibara H, Nimura Y. Doppler echocardiographic features of coronary arteriovenous fistula: complementary roles of cross sectional echocardiography and the Doppler technique. *Br Heart J* 1984;51:508-18.
18. Cooper MJ, Bernstein D, Silverman NH. Recognition of left coronary artery fistula to the left and right ventricles by contrast echocardiography. *J Am Coll Cardiol* 1985;6:923-6.
19. Ludomirsky A, Danford DA, Glasow PF, Blumenschein SD, Murphy DJ, Huhta JC. Evaluation of coronary artery fistula by color-flow Doppler echocardiography. *Echocardiography* 1987;4:383-6.
20. Shah RM, Nanda NC, Hsiung MC, Moos S, Roitman D. Identification of anomalous origin of the right coronary artery from the pulmonary trunk by Doppler color flow mapping. *Am J Cardiol* 1986;57:366-7.